A very uncommon duplicity
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Section: Genital (female) imaging
Area of Interest: Genital / Reproductive system female
Urinary Tract / Bladder Colon
Procedure: Diagnostic procedure
Imaging Technique: MR
Imaging Technique: Ultrasound
Special Focus: Congenital Case Type: Clinical Cases
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Patient: 30 years, female

Clinical History:
A 30-year-old female patient was referred to the gynaecology department for suspected pelvic inflammatory disease. In the gynaecological examination, the external genitalia were normal, but in the vaginal introitus two vaginas are identified. Transvaginal ultrasound confirmed the presence of a double uterus. Pelvic inflammatory disease was discarded and magnetic resonance imaging (MRI) was recommended.

Imaging Findings:
An abdominal ultrasound showed double bladder and uterus (Fig. 1). MRI was performed to adequately characterize the findings and the associated anomalies. Imaging Findings: The findings were (Fig. 2-4): Presence of double uterus and cervix with duplication of the vagina (Fig. 2). Urinary system: two bladders with double ureteropelvic systems in both kidneys (Fig. 3). Gastrointestinal tract: colorectal duplication with two ani (Fig. 2 and 4).

Discussion:
Caudal duplication syndrome (CDS) is a rare entity with prevalence of less than 1 per 100.000 at birth [1] and there are no consistent risk factors or familial associations [2]. Only about 40 cases have been reported in the literature [3]. It was first described by Dominguez et al. in 1993. They described six children with duplications of distal organs derived from the hindgut, neural tube, and adjacent mesoderm and they coined the term CDS to describe this association [4]. Common gastrointestinal anomalies include duplication of the colon and rectum, which may be associated with a variety of other anomalies such as imperforate anus, rectal fistula, ventral hernia, omphalocele, duplication of terminal ileum, double appendices, Meckel's diverticulum, intestinal malrotation, and situs inversus. Urogenital anomalies comprise duplication of the external and internal genitalia, ureters and bladder and anomalies of the kidney [5]. It is also associated to spinal dysraphism, kyphoscoliosis of the spine and vertebral body defects [6, 7]. In girls, two uterus and double vagina may be seen; in males, double phalluses.

Although most reported cases have some of these features, our patient has almost all of them (including spina bifida and kyphoscoliosis as associated abnormalities), except for abdominal wall defects. It is very uncommon to find a patient with these combination of abnormalities.

The exact aetiology of caudal duplication syndrome is unknown. There are several theories, but none has been
universally accepted. Dominguez et al proposed that it may result from an insult to the caudal cell mass and hindgut between 23rd-25th days of gestation [4]. Some authors suggest that it may result from an early insult to the urorectal septum [1] or from an incomplete division of monozygotic twins [5, 8]. It has also been postulated that the disorder is related to misexpression of one or more of the distal HOX genes, potentially HOX10 or HOX11, leading to abnormal proliferation of caudal mesenchyme [1, 8].

The detection of such abnormalities is important for several reasons. In females, it may cause problems in sexual intercourse and pregnancy complications [9].

Many of these anomalies are initially diagnosed at hysterosalpingography (during the study of infertility or recurrent pregnancy loss) or by ultrasound. MRI has high accuracy in the evaluation of Müllerian duct anomalies, genitourinary and gastrointestinal malformations [9].

Treatment in these patients might be individualized because of the many associated anomalies. Surgical procedures may be required depending on the multiple anatomic variables [1].

**Differential Diagnosis List:** Caudal duplication syndrome., Simple failure of Müllerian fusion, Persistent cloaca

**Final Diagnosis:** Caudal duplication syndrome.

**References:**


Description: Transabdominal ultrasound image of double bladder (B) and uterus (U). Origin: Department of Radiology, H. Universitario Dr. Peset, Valencia, Spain
Figure 2

Description: T2 TSE coronal and axial images show double uterus (asterisks), duplication of vagina (arrowheads) and anus (arrows). Origin: Department of Radiology, H. Universitario Dr. Peset, Valencia, Spain.
**Description:** T2 TSE coronal and axial images show double bilateral ureteropelvic systems (arrowheads) and two bladders (asterisks). **Origin:** Department of Radiology, H. Universitario Dr. Peset, Valencia, Spain.
**Description:** The image on the left shows a splenic flexure of the colon (red arrow) and the right shows the splenic flexure of the doubled colon (white arrow). **Origin:** Department of Radiology, H. Universitario Dr. Peset, Valencia, Spain.